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# Eosinophilic Granulomatosis with Polyangitis – Case Report

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## Introduction

EGPA is an uncommon small-medium vessel vasculitis. The presentation varies from asymptomatic to severe clinical manifestations including cardiovascular, gastrointestinal, renal and cns involvement.

Most common involvement is respiratory followed by dermatological manifestations

## Case Report

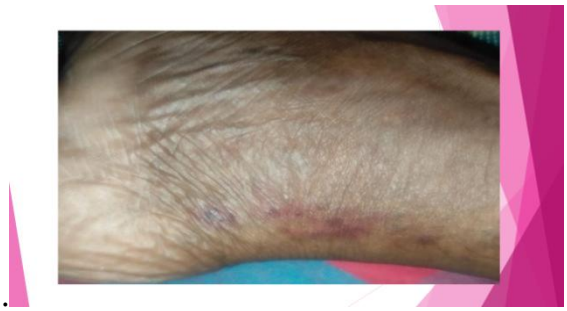
52/M patient with history of asthma from childhood, on regular medication, now presented with short febrile illness and acute onset right wrist drop, followed by development of left foot drop 3 days later.

Patient also gives history of positive sensory symptoms involving distribution of right radial nerve and B/L common peroneal nerve.

No history of proximal muscle weakness, no history suggestive of higher function or cranial nerve involvement. No history of back pain or autonomic symptoms.

On examination PR 78/' regular, BP 140/80, no postural hypotension, Temperature 101F.

There was b/l pitting pedal oedema. Palpable purpura was noted on right lower limb above medial malleolus



On central nervous system examination, there was weakness of extension of right wrist and fingers and weakness of dorsiflexion of left ankle and decreased sensation along the distribution of right radial nerve and bilateral common peroneal nerve.

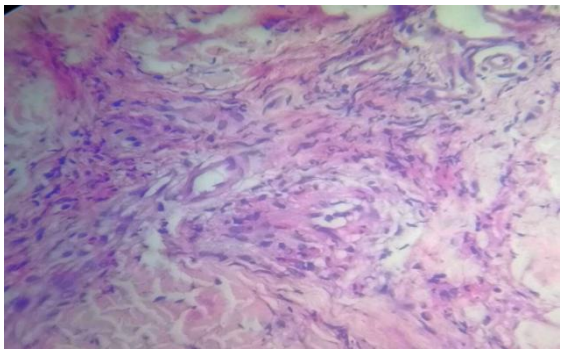
On auscultation, Bilateral rhonchi was present.

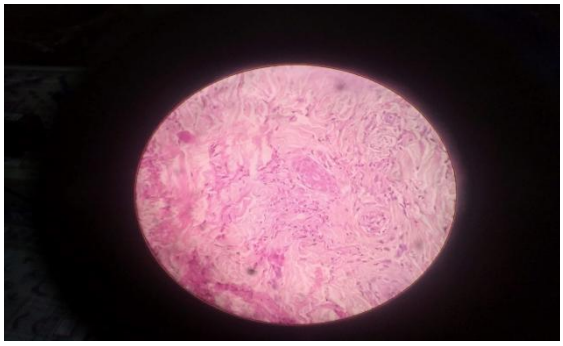
All other systemic examinations were normal.

Clinically we suspect the possibility of a confluent type of mononeuropathy multiplex in view of involvement of more than two peripheral nerves and was investigated for the cause.

Investigations showed Hb 14.5, ESR 53, Total count 15800, Platelet count 2.72L Absolute

eosinophil count 7740, IgE 1593, CRP positive, C-ANCA, P-ANCA negative, Peripheral smear showed severe eosinophilia. N37E50L3
Eosinophils 9000/mm3
ECG: Normal sinus rhythm, 2D ECHO: Normal
CXR: B/L reticulonodular calcifications were present.
USG Abdomen: Hepatomegaly
VDRL, HIV, HBSAg, Anti HCV were negative, Tumour markers, URE, RFT, LFT, RBS, TFT, FBS, FLP, RBS, PT, INR, APTT, BT, CT, ANA Profile were within normal limits.
Video bronchoscopy, BAL, split skin smear, MRI brain and whole spine screening showed no significant findings.
Skin biopsy from purpura showed Eosinophilic vasculitis with fibrinoid necrosis and tissueeosinophilia.





We arrived at the diagnosis of Eosinophilic granulomatosis with polyangitis according to 2022 American college of Rheumatology / European alliance of association for rheumatology classification criteria for EGPA.
Patient was started on pulse steroid therapy.

**Discussion**

EGPA is an uncommon disease with an estimated annual incidence of 1-3 per million
The mean age of onset is 48 years, with female to male ratio of 1.2:1. It involves small – medium sized muscular arteries, capillaries, veins and venules. A characteristic histopathological feature is granuloma that may be present in tissues or within the walls of vessels themselves., associated with infiltration of the tissue with eosinophils. Precise pathogenesis of disease is uncertain and it points to aberrant immunologic phenomena.
Patients with EGPA often exhibit non-specific manifestations like fever, malaise, anorexia and weight loss. Severe asthmatic attacks and pulmonary infiltrates dominate the clinical picture. Mononeuritis multiplex is the second most common manifestation and occurs in 72% patients. Skin manifestations occur in 51% patients and include purpura in addition to cutaneous and subcutaneous nodules. Myocarditis, pericarditis, endocarditis or coronary vasculitis can occur in 14% of patients and is an important cause of mortality.
The characteristic laboratory findings include striking eosinophilia >1000cells/ ul.in >80% people. Elevated ESR, CRP, about 48% have circulating ANCA. Although diagnosis of EGPA is optimally made by biopsy in a patient with evidence of asthma, peripheral blood eosinophilia, and clinical features consistent with vasculitis.
Treatment include glucocorticoids, dosetapering is often limited by asthma and many patients require low dose prednisolone for persistent asthma, even after clinical recovery from vasculitis. In patient who present with fulminant multi system disease, particularly cardiac involvement the treatment of choice is a combined regimen of daily cyclophosphamide and prednisolone followed by azathioprine or methotrexate.
Our patient had confluent type of presentation with involvement of more than one nerve, within a short time. It is an extremely rare presentation of mononeuropathy multiplex due to EGPA.
The prognosis of untreated EGPA is poor with reported 5-year survival of 25%. With treatment,

prognosis is favourable, hence timely diagnosis and proper management is essential for a favourable prognosis.

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CLASSIFICATION CRITERIA FOR EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- These classification criteria should be applied to classify a patient as having eosinophilic granulomatosis with polyangiitis when a diagnosis of small- or medium-vessel vasculitis has been made
- Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria

CLINICAL CRITERIA

| | |
|---|---|
| Obstructive airway disease | +3 |
| Nasal polyps | +3 |
| Mononeuritis multiplex | +1 |

LABORATORY AND BIOPSY CRITERIA

| | |
|---|---|
| Blood eosinophil count ≥ 1 x10⁹/liter | +5 |
| Extravascular eosinophilic-predominant inflammation on biopsy | +2 |
| Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies | -3 |
| Hematuria | -1 |

Sum the scores for 7 items, if present. A score of ≥ 6 is needed for classification of EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS